

### **Appendix 1. Risk Indicators Associated With Permanent Congenital, Delayed-Onset, or Progressive Hearing Loss in Childhood.**

1. Caregiver concern\* regarding hearing, speech, language, or developmental delay (Roizen, 1999).
2. Family history\* of permanent childhood hearing loss (Cone-Wesson et al., 2000; Morton & Nance, 2006).
3. Neonatal intensive care of >5 days, or any of the following regardless of length of stay: ECMO,\* assisted ventilation, exposure to ototoxic medications (gentamycin and tobramycin) or loop diuretics (furosemide/lasix), and hyperbilirubinemia requiring exchange transfusion (Fligor et al., 2005; Roizen, 2003).
4. In-utero infections, such as CMV,\* herpes, rubella, syphilis, and toxoplasmosis (Fligor et al., 2005; Fowler et al., 1992; Madden et al., 2005; Nance et al., 2006; Pass et al., 2006; Rivera et al., 2002).
5. Craniofacial anomalies, including those involving the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies (Cone-Wesson et al., 2000).
6. Physical findings, such as white forelock, associated with a syndrome known to include a sensorineural or permanent conductive hearing loss (Cone-Wesson et al., 2000).
7. Syndromes associated with hearing loss or progressive or late-onset hearing loss,\* such as neurofibromatosis, osteopetrosis, and Usher syndrome (Roizen, 2003). Other frequently identified syndromes include Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson (Nance, 2003).
8. Neurodegenerative disorders,\* such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome (Roizen, 2003).
9. Culture-positive postnatal infections associated with sensorineural hearing loss,\* including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis (Arditi et al., 1998; Bess, 1982; Biernath et al., 2006; Roizen, 2003).
10. Head trauma, especially basal skull/temporal bone fracture\* requiring hospitalization (Lew et al., 2004; Vartialnen et al., 1985; Zimmerman et al., 1993).
11. Chemotherapy\* (Bertolini et al., 2004).

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\* Risk indicators that are marked with an asterisk are of greater concern for delayed-onset hearing loss.